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Teaching NeuroImage: Cerebellar Atrophy Due to JC Virus Granule Cell Neuronopathy: A Clinical Syndrome Distinct From Classic PML

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Contributions:

Carlos Silva-Rosas: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

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A 34-year-old Hispanic man with HIV infection on no treatment (CD4 $20/\mu l$, viral load $1.9/10^6$ copies/ml) presented with four months of dizziness, ataxia and scanning speech consistent with a pancerebellar syndrome.

MRI scan (Figure) demonstrated marked cerebellar atrophy. CSF showed normal cell count, protein and glucose levels, non-reactive VDRL. CSF PCR was negative for cytomegalovirus, varicella-zoster, herpes simplex type 1 and 2, Epstein-Barr, and herpes virus 6, but positive for JC virus. JC virus granule cell neuronopathy (GCN) was diagnosed. JC virus variants may rarely infect cerebellar granule neurons instead of oligodendrocytes as seen in classic PML with white matter involvement. HAART (zidovudine, lamivudine, efavirenz) commenced immediately, with slight symptomatic improvement at 12 months, MRI scan was unchanged.

Clinicians should suspect JC virus strain infection producing GCN in AIDS patients with symptomatic cerebellar atrophy ² and commence HAART promptly- IRIS is not usually a concern in cases of isolated JCV GCN.

http://links.lww.com/WNL/C655

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Figure (A) Sagittal T1-weighted brain MRI and (B) Axial FLAIR-weighted brain MRI show marked cerebellar atrophy (arrows)





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